LEPROSY SIMULATING SYRINGOMYELIA*

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Some of the members present may remember a woman sent to the City and County Hospital about two years ago with the diagnosis "leprosy." Her hands showed extreme trophic changes and mutilations of the Morvan type; there were marked disturbances of sensation to pain and temperature and advanced atrophy of small hand muscles and muscles of the shoulder girdle. The presence of bulbar symptoms, however, of marked scoliosis, bladder symptoms, extreme spasticity of the lower extremities with clonus and Babinski decided absolutely for syringomyelia and against leprosy.

This boy of eleven, born in the Cape Verde Islands, of Portuguese parents, has been under observation in hospital during the past weeks. His history is difficult to obtain, as his mother refers all symptoms to falls at different times. The father is living in the Cape Verde Islands, and has had some sort of a "sore" on one leg for years. The beginning of the affection is variously dated. The right foot has slowly assumed its present position during the past five or six years. The affection of the left hand was first noticed after a fall on the ice in New York, December, 1903, and the hand was thought to have been frozen. There never has been pain. With this unsatisfactory history we come to the examination:—

The skin in irregular areas over the neck, trunk, arms and thighs is a muddy yellowish brown. Over the forehead it looks thin, and contains less than the normal amount of pigment. Scattered over the trunk, arms and thighs are circular or oval whitish patches, in some places on the back almost exactly symmetrical. These patches are not anesthetic, and sweat equally with other parts of the skin after pilocarpin administration.

There are numerous small hard glands in the posterior triangles of the neck, in the groin and axillæ. The ears are not deformed. Teeth are normal. The nose is thick and somewhat flattened. There is no evidence of septum ulceration. There are no stigmata of syphilis. The face is irregular, but there is no sympathetic lesion.

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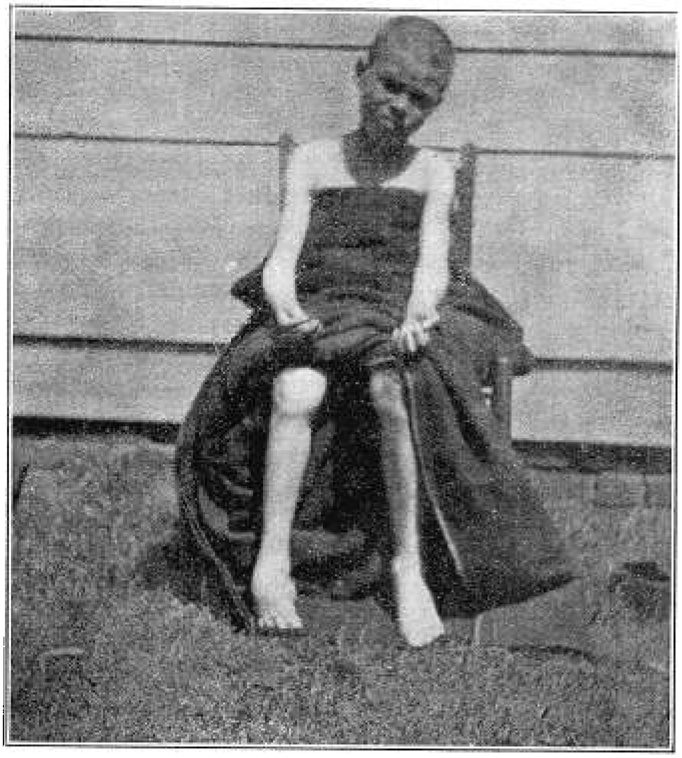
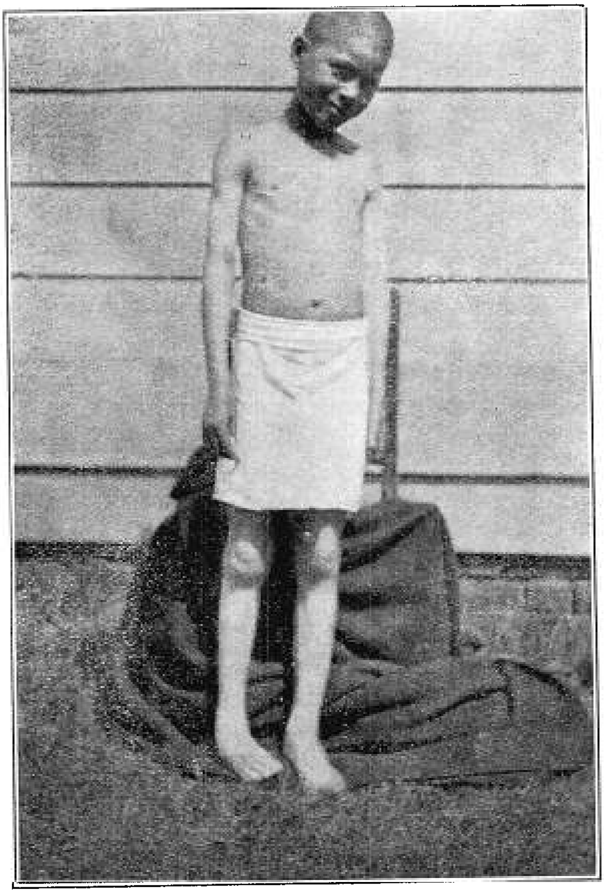
Pupils are normal. Nystagmus is absent. Cranial nerves are unaffected.

The left small occipital, the right great auricular and both ulnar nerves above the elbow are decidedly thickened. The right ulnar nerve is tender. There is no scoliosis.



The left hand is a typical claw-hand with clenched fingers and atrophy of the small muscles. Trophic changes are shown by the deep cracks, thickened skin and nails, small ulcers, and deep purplish color. There is reaction of degeneration in the small hand muscles. The fingers can be extended at the first phalanx only; the thumb cannot be fully extended or abducted.

There is no rigidity of arm or forearm, and arm reflexes are normal. There are beginning trophic changes in the right hand, but no muscle atrophy. There is no palsy except of atrophied muscles.



Abdominal muscles and reflexes are normal.

There is no bladder disturbance.

The right foot is in marked equino-varus position. Two perforating ulcers on the outer border mark the points of pressure. On the heel is a deep, fresh wound caused by a nail in his shoe which projects fully half an inch, but it caused no pain and was not noticed.

The nails are curved and thickened.

There is no palsy except in the atrophic group of foot extensors. The tibialis anticus and peronei give complete R. D. There is absolute lack of rigidity. Knee and left Achilles jerks are normal. The right Achilles cannot be obtained. There is no Babinski. The plantar reflex is abolished. Romberg and ataxia are not present.

Sensory findings vary somewhat on different examinations, and there seems now a functional addition to the original changes.

There is loss of sensation to temperature in the whole of the left ear. There is thermanesthesia down a strip about 1½ inches wide on the posterior surface of the left arm. Below the elbow there is complete analgesia and thermanesthesia with definite hypesthesia. The prick of a pin is felt as a touch along the radial border of the arm and on the thumb and first finger, but not along the ulnar side.

In the right forearm analgesia and thermanesthesia are definitely more marked in the distribution of the ulnar nerve.

Deep sensation is approximately normal. There are no sensory changes over the trunk or thighs.

In the right leg the areas of athermia, analgesia and anesthesia cover the entire foot and the lower half of the leg, running up higher on the outer and posterior surfaces.

On the left side there is anesthesia, analgesia, and thermanesthesia over the sole, a small patch on the outer side of the calf, over the fourth and fifth toes, and over the dorsal surface of the leg to the middle of the calf. X-Ray shows the foot bones to be normal.

The appearance of the foot might suggest the neural form of progressive muscle atrophy, but the affection is usually symmetrical, of slower development, and such extensive sensory changes do not occur. The affection described by Dejerine and Thomas under the head of "Névrite interstitielle hypertrophique et progressive de l'enfance" is marked by thickening of peripheral nerves, but pain, ataxia, changes in reflexes are common features.

Differential diagnosis has to consider essentially two affections—syringomyelia and leprosy. Against syringomyelia would speak:—

(1) The absence of sympathetic eye signs in presence of the involvement of the small muscles of the hand supplied by the first dorsal segment; (2) the absence of scoliosis; (3) the absence of rigidity and of increased reflexes: especially the absence of these signs in the lower extremities; (4) the distribution of the palsies—left hand and right foot, and the limitation of palsies and atrophies to the distal parts of the extremities; (5) the absence of ataxia and sphincter involvement; (6) the loss of temperature sense in the left ear, and the limitation of sensory changes in the right arm to the areas supplied by the ulnar nerve; (7) the absence of Babinski and the abolition of the plantar reflex; (8) the widespread involvement of sensation in the lower extremities; (9) the thickening of the peripheral nerves.

The skin changes would weigh more for leprosy. Leprosy, as we know, is common in the Cape Verde Islands. Bacilli have been sought in the blood, in nasal secretion, and in blister fluid without result.

The boy will be given Shaulmoogra oil and, later on, Calmette's serum.